THREE CASES OF THE FAMILY TYPE OF CEREBRAL DIPLEGIA.

BY F. X. DERCUM.

The following cases, because of their rarity and because of the family history, are exceedingly interesting and worthy of record. They are all children. The father presents a family history which is negative as regards nervous and mental affections. The mother of the children is his third wife. He had also had a number of children by both his first and second wives; all of these children were normal. The third wife has borne him four children, three of whom present marked spastic diplegia. The family history of the third wife, the mother of the diplegic children, is interesting from the fact that the child of one of her paternal aunts is paralyzed—she thinks in a manner similar to that of her own children. She herself is perfectly healthy. Every one of her four labors was normal. No delay or incident worthy of mention occurred. The history of the children is briefly as follows:

Oscar, eleven years of age, presents the symptoms of a typical spastic diplegia. He was normal at birth and continued well up to sixteen months of age. He was able to walk unassisted, his gait seemed perfectly normal, and he was able to say quite a number of words and understand much that was said to him. At this time he was seized with a convulsion which was general, and during which he was unconscious. After the convulsions he seemed stupid, had lost largely the power of speech, and was weak and helpless. Some time afterward it was noticed that his legs were stiff, and when he began to use them, it was seen that he moved them very awkwardly. A similar condition of affairs was present in the arms. The spastic symptoms seem to have made their appearance gradually, and finally became pronounced.

At present he is able to walk, but can do so unsupported for a distance of a few feet only. The gait is spastic and tottering. The legs are quite rigid in extension. The arms are also spastic, but to a much less degree than the legs. Athetoid movements of the hands are now and then observed. When seated the feet assume the position of talipes equino-varus. His speech is so defective that he can speak only two or three words, and these are uttered so indistinctly that they can be comprehended by his parents only. Subsequent to the convulsion he had attacks of *petit mal* during which his head would suddenly drop. At times he would fall forward. His head is much bruised from falls received during these attacks. He is exceedingly stupid and quite idiotic; he is also incontinent.

Justus, six years, was well until four years of age. Up to this period he had been a bright and apparently normally developed child. At this time he was attacked by measles, the attack appearing, however, to be one of moderate severity. During the convalescence a profound change was noted in him. He was stupid, and appeared to be weak in both arms and legs, the weakness being most profound upon the right side.

Justus presents an excessively marked spastic diplegia. Both legs are exceedingly rigid, the legs being slightly flexed at the knees, the feet completely extended. The right arm is held firmly in a semi-flexed position. The left arm is also spastic, but the patient is able to move it to some extent. He frequently attempts to walk, but invariably falls to the ground; and while in this position he writhes with choreiform and athetoid movements. At other times, when sitting in his father's lap, spasmodic movements are observed in legs, arms and face. He is also epileptic, his seizures having the character of *petit mal.* His intelligence appears to be somewhat better than that of Oscar; he talks a little more, and the words are uttered more distinctly, although bradylalia is pronounced. He is not entirely continent, sometimes not telling his mother when his bladder or bowels are to be emptied.

Cleveland, two years old, was a normal child at birth and appeared to develop in a perfectly normal manner until he was two years of age. He was then attacked by measles at the same time as his brother Justus and, like Justus, during convalescence became diplegic and epileptic. During one of his early epileptic attacks he appeared to loose his vision, but this subsequently returned. At present he has marked diplegia, but is able to walk without falling. He can, although with difficulty, pick up small objects from the floor. Athetoid movements are present in the fingers as in the other cases. He appears also to be somewhat more intelligent than his brothers.

All of the children present tumid, sodden features like those of confirmed epileptics. Oscar and Justus look stupid and idiotic, though this cannot be said of Cleveland. In all of the cases the reflexes are exaggerated wherever the contractures and rigidity do not interfere with their being studied. In none of the cases was there nystagmus. Owing to the restlessness and poor mental condition of the children it is impossible to make a satisfactory examination of the eye ground. Vision appears, however, in all three to be somewhat defective. All of the cases drool more or less, especially is this true of Cleveland.

An examination of the remaining child, the second in age, a boy of nine and a half years, proved negative. He is in average physical and mental health, and his intelligence is fully up to the average, if not above. It was interesting to note that this child was not attacked by measles, nor has he had any other serious illness.

In these cases there seems to be present a feebleness of development on the part of the neurons of the motor area. It would seem that if not interfered with, they had the power of developing in a normal manner but that their power of resistance was so low that they underwent degenerative changes from very slight causes. In two of the cases the cause seems to have been the toxicity of measles. Freud regards these cases of family diplegia as analogues of Friedreich's ataxia, and in a sense this is true.

Cases of diplegia of the family type are very rare. A number of cases, however, are on record, among which are the following:

Pelizeaus (Archiv f. Psychiatrie, Vol. XVI., 1895) describes five cases of the affection occurring in three generations. All of these cases were males and presented in addition to spastic diplegia, imbecility. Gee (St. Bartholomew's Hospital Reports, 1889, XXV., p. 81) describes three cases. His cases are remarkable from the fact that they instance transmission of the disease through the father. The first case was thirty-seven years of age, and the father of the other two. His symptoms were those of spastic diplegia. His family history presented the following features: a maternal aunt completely paralyzed, two maternal cousins who were born deaf and dumb, and one cousin who was born without fingers. His daughter, aged twelve, and his son, aged eleven, constitute the second and third cases described by Gee, and presented typical spastic diplegia. Sachs (Jour. of Nervous and Mental Disease, 1887 and 1882) reports two instances of diplegia occurring in families. First, two sisters, one of them becoming diplegic at the age of three months, and the other at the age of eight months; the former presenting spastic palsy and the latter flaccid palsy. In the second instance there were four children who after a period of normal development became spastic and idiotic. Massolongo (cited by Freud) records three sisters, who each at the age of seven years developed spastic paralysis and athetosis. Freud (Neurolog. Zentralbl., 1893, XII., pp. 512 and 542) places on record the following interesting cases. A physician had married his niece, the child of his sister. Six children were born to this couple. The first was born prematurely and died soon after birth. The second and third presented spastic diplegia. The fourth child was normal. The fifth died paralyzed and idiotic. (This child Freud did not himself see.) The sixth child was born prematurely and also died early. The three children which survived were all boys. One of them was normal whilst the other two had spastic diplegia. In addition, the latter presented atrophy of the optic nerves, nystagmus and bradylalia.

DEATH IN HYSTERIA.

M. R. Le Fournier (Thése de Paris, 1896) writes upon the possibility of death in hysterical troubles, and maintains that hysteria should not be regarded as a disease having always a favorable prognosis. Death sometimes takes place from violent convulsive attacks with spasm of the glottis. Anorexia occasionally causes death by inanition, and the author lays some stress on the danger of hypnotization without proper precautions, and points out that it has sometimes been known to bring on fatal spasmodic attacks. MITCHELL